As indicated by Flynt, in an article in this issue, birth defects have taken a primary place within the spectrum of pediatric conditions and will increasingly demand clinical attention. The impact of survival with varying degrees of handicap has wide implications for the affected child, the family, and the community.

In our current era of world population pressures, questions have been raised regarding the desirability of encouraging survival of persons with physical, mental, metabolic, or other limitations.

Nowhere is this question more pertinent today than in the case of meningomyelocele. The condition occurs in one to two per 1,000 live births in the U.S.\(^1\,\,^2\) Its highest reported inci-

---

By ALFRED L. SCHERZER, Ed.D., M.D.
idence is in Western England, Wales, and Ireland (three to four per 1,000 live births). \textsuperscript{3-1,5} Isolated pockets of high incidence are also noted in Alexandria, Egypt, and in the Punjab region of India. \textsuperscript{6}

The condition occurs more frequently in girls, is more common among Catholics, and is rarely found among Jews. An inverse relationship has been noted between the condition and socioeconomic status; maternal age and birth rank have not been found to be significantly related. \textsuperscript{7} A definite familial tendency is noted—risk of recurrence is about six per cent for subsequent children if there is one previously affected sibling and 10 per cent or greater if there are more affected children in the family. \textsuperscript{4}

Meningomyelocele is a spinal defect with failure of appropriate neural tube closure. It is present by the 25th to 29th day of gestation. \textsuperscript{9} Thus, the defect is frequently present before a definite pregnancy is confirmed. It is most common in the lumbar or lumbosacral area (Figure 1). Clinical deficits include: 1. Flaccid paraplegia below the level of the lesion; 2. Neurogenic bladder; 3. Neurogenic bowel; 4. Orthopedic problems; 5. Hydrocephalus. The hydrocephalus is generally due to an Arnold-Chiari anomaly and is present in 60 to 80 per cent of cases.

Etiologic studies to date have been unrevealing. Most recently, attention has been given to possible statistical relationships to ingestion of blighted potatoes. \textsuperscript{10} However, no confirmatory evidence is currently available. From the information now known it is generally felt that the disorder is probably polygenic in origin with unknown contributory environmental factors.

Fifteen years ago the outlook for

\textit{continued}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{image.png}
\caption{Typical lumbar level meningomyelocele}
\end{figure}

\textbf{Etiology of meningomyelocele is unclear}

\textbf{Dr. Scherzer} is associate professor of pediatrics, Cornell University Medical Center; director, Division of Pediatric Habilitation Services, New York Hospital, New York, N.Y.; chief of medical services, Nassau County Cerebral Palsy Treatment and Rehabilitation Center; and consultant, Office of Special Education, New York City Board of Education, and Bureau for Handicapped Children, New York City Department of Health.
survival of children with meningo-myelocele was bleak. At least 38 per cent died within the first month and close to 100 per cent were dead by one year. Both surgical and medical advances have subsequently changed this picture considerably (Table 1).

Closure of the back within 12 to 24 hours has been shown to both reduce infection and preserve neural function. Treatment of hydrocephalus has become more effective as atrial and particularly peritoneal shunts have been better designed. Careful and continuous monitoring of urinary infection has been followed with appropriate antibiotic management. Above all, the ileal conduit has become available for diversion at the earliest signs of reflux or upper tract changes, thus eliminating renal failure as a later cause of morbidity. Bracing, physical therapy, and orthopedic procedures have enabled a surprising degree of physical independence in many of these children.

Centers which have used these modalities at the appropriate time have shown a complete reversal in mortality with a survival rate exceeding 70 to 80 per cent. Increasing numbers of these children are now coming into the teenage years with varying degrees of function.

Several studies find encouragement in the long-term results of a multi-specialty approach. In a group of 171 Philadelphia children, 42 per cent had I.Q.’s of 80 or better and were ambulatory with or without support. At New York Hospital, an emerging school-age population showed an average I.Q. of 82; 13 of the 16 children were ambulatory and 15 were in either regular or special day-school programs.

Other centers show similar results and we are now being faced with increasing numbers of survivors who are striving to better understand the meaning and usefulness of their own existence.

The expense of long-term habilitation management has been estimated at from $250,000 to $300,000 per patient over many years of treatment. The cost in terms of emotional upheaval of the family, adjustment of the child, and special school and vocational facilities is more difficult to calculate. On the other hand, the present and future contribution to society of the functional survivors has yet to be measured.

In spite of obvious gains in mortality and morbidity, some people continue to seriously question the advisability of full management of this condition on medical, financial, or ethical grounds. House staff physicians, in particular, frequently have a negative attitude, which is most likely the result of limited experience with the level of function possible with these children.

A recent review by Lorber has stirred particular interest. He reviewed two series of patients given "optimum" medical and surgical treatment

---

**TABLE 1**

**MENINGOMYELOCELE**

"Standard" Procedures of Management

1. Closing of sac within 24 hours
2. Shunting for hydrocephalus: observation for malfunction
3. Treatment of urinary infections
4. Regular urology evaluation—IVP, cystogram
5. Ileal conduit on medical indication
6. Physical therapy for motor development
7. Orthopedic surgery for correction of deformities
8. Social service support and guidance
9. Educational and social development guidance
10. General pediatric care

---

Full management is being questioned on medical, financial, or ethical grounds.
Who should decide who has the right to live—how often are parents listened to?

facilities for custodial care are generally minimal and frequently non-existent.

What of the parents? Their voices are heard little. And how often are they heeded? The titles of two recent articles in consumer magazines—"The Agonizing Decision of Joan and Roger Pell" and "I Left My Newborn Son to Die"—give some indication of the indecision and doubt that surround the presence of a deformed child.

A recent provocative article discussed prospects for a hospital review board responsible for decisions regarding treatment—"The God Committee." Professional reactions to this concept are as varied as the individual personalities involved.

Parents' reactions to "The God Committee" idea were recently obtained by Swinyard through the use of a questionnaire involving 1,300 children. Preliminary results indicate that 80 per cent of the parents would totally reject such an idea and about 80 per cent feel that the medical profession has an obligation to provide maximum care, at least until such time as preventive measures are available.

This information is at variance with the opinion of Dr. Lorber, obtained from colleagues, that "... many parents express the wish that their child should not be treated if it will mean permanent handicap with major disabilities, provided also that their child's life is not unduly prolonged."

These issues are now becoming more apparent as medical and surgical procedures literally enable choices to be made. A birth defect such as meningomyelocele becomes at once a model for multidiscipline management and a prime case for ethical considerations in medicine. Everyday, decisions regarding treatment of such conditions are increasingly becoming the concern of medical institutions in this country.
At Harvard, for example, an Inter-
faculty Program on Medical Ethics has
been established, and "ethics" rounds
are being held at the Children's Hos-
pital Medical Center in Boston involv-
ing medical specialists, clergymen,
and lawyers. Similar programs are be-
ing established at Georgetown, Co-
lumbia, and Dartmouth. A recent sur-
vey by the Institute of Society, Ethics,
and the Life Sciences reports, how-
ever, that only 17 medical schools
have an identifiable program, al-
though 33 have some elective cours-
es.54

The issues in ethical medicine are
complex, and the attitudes of profes-
sionals, patients, families, and the
community need as careful study as
other facets of medical management.
The time has come for centers of
health care to establish the climate,
organization, and framework around
which these questions of ethical con-
sideration can be dealt with openly in
a manner acceptable to all concerned.

More data are needed regarding the
natural history and long-term conse-
quencies of those who have benefited
from present medical management. A
better appreciation of how the phy-
sician can become more objective in his
or her approach to the issues is es-
sential.

By raising these questions now,
birth defects are demanding solutions
today to much of medicine for the
future. For it is clear that emphasis is
changing from acute care to long-term
management. And, increasingly, the
physician will have to join others in
the critical decisions not only of life
or death, but of the quality of sur-

BIBLIOGRAPHY

1. Alter, M. Anencephalus, hydrocephalus, and spin
a bifida. Arch. Neurol. 7 (1962), 411.
84 (1952), 35.
3. Record, R. and McKeown, T. Congenital
malformations of the central nervous system. I.
183.
4. Laurence, K. The survival of untreated spina
5. Stevenson, A. and Warnock, H. Observations
on the results of pregnancies in women resident
in Baltim. J. Data relating to all pregnancies end-
6. Stevenson, A. et al. Congenital malforma-
tions: Report of study of series of consecutive
births in 24 countries. Bull. W.H.O., Suppl. 34
(1966), 1.
7. Naggar, L. and MacMahon, B. Ethnic differ-
ces in the prevalence of anencephaly and
277 (1967), 119.
8. Carter, C. Spina bifida and anencephaly—
a problem in genetic-environmental interaction.
9. Patten, B. Embryological stages in the es-
ablishing of myeloschisis with spinabifida. Am.
J. Anat. 93 (1953), 365.
10. Renwick, J. Hypothesis: anencephaly and
spinabifida are usually preventable by avoid-
ance of a specific but unidentified substance
11. Rickham, P. and Mawdsley, T. The effect
of early operation on the survival of spinabifida
(1966), 20.
12. Sharrard, W. et al. A controlled trial of
immediate and delayed closure of spina bifida
13. Shulman, K. and Ames, M. Intensive treat-
ment of 50 children born with meningomyeleoce.
14. Ames, M. and Schut, L. Results of treat-
ment of 171 consecutive meningomyeleoces—
15. Schenzer, A. and Gardner, G. Studies of the
school-age child with meningomyeloce. 1.
Physical and intellectual development. Pediatrics
16. Andrews, L. Spina bifida cystica—a follow-
17. Let blighted babies die, or not? Med. World
News 13 (Nov. 17, 1972), 27.
18. Bunch, W. et al. Modern Management of
Myelomeningoce. St. Louis: Warren H. Green,
1972.
19. Lorber, J. Results of treatment of meningo-
279.
20. Pell, R. The agonizing decision of Joan and
21. Rotter, W. I left my newborn son to die.
22. Freeman, E. The God Committee. New
24. Ethical questions Hippocrates did not
have to face. Med. World News 13 (July 14, 1972),
41.